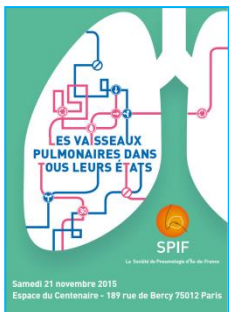


# Hypertension pulmonaire à l'échocardiographie: que faire?



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## 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

**The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)**

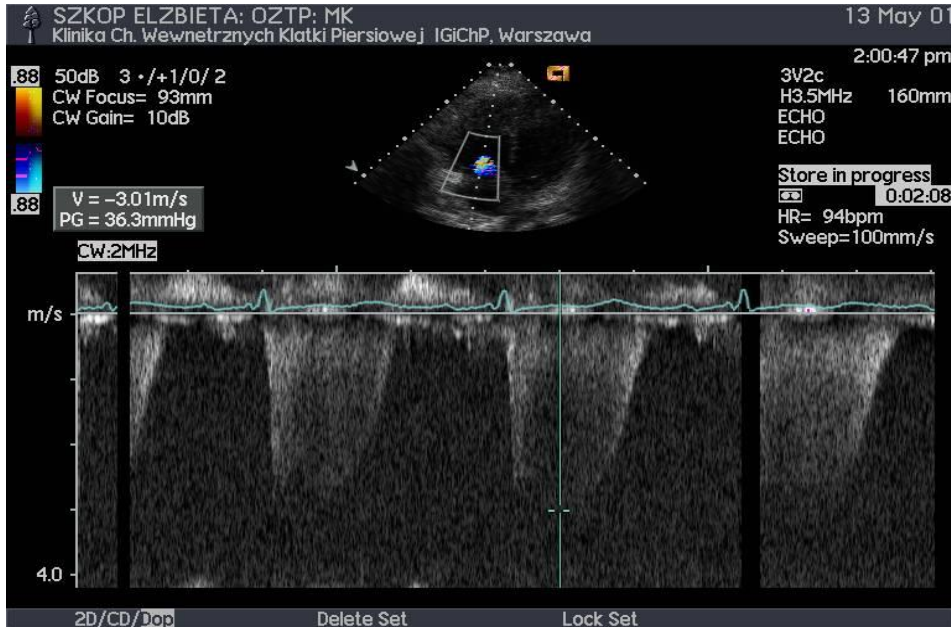
**Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)**

**Authors/Task Force Members: Nazzareno Galiè\* (ESC Chairperson) (Italy), Marc Humbert\*<sup>a</sup> (ERS Chairperson) (France), Jean-Luc Vachiery<sup>c</sup> (Belgium), Simon Gibbs (UK), Irene Lang (Austria), Adam Torbicki (Poland), Gérald Simonneau<sup>a</sup> (France), Andrew Peacock<sup>a</sup> (UK), Anton Vonk Noordegraaf<sup>a</sup> (The Netherlands), Maurice Beghetti<sup>b</sup> (Switzerland), Ardeschir Ghofrani<sup>a</sup> (Germany), Miguel Angel Gomez Sanchez (Spain), Georg Hansmann<sup>b</sup> (Germany), Walter Klepetko<sup>c</sup> (Austria), Patrizio Lancellotti (Belgium), Marco Matucci<sup>d</sup> (Italy), Theresa McDonagh (UK), Luc A. Pierard (Belgium), Pedro T. Trindade (Switzerland), Maurizio Zompatori<sup>e</sup> (Italy) and Marius Hoeser<sup>a</sup> (Germany)**



# Measurement of Pulmonary arterial pressure

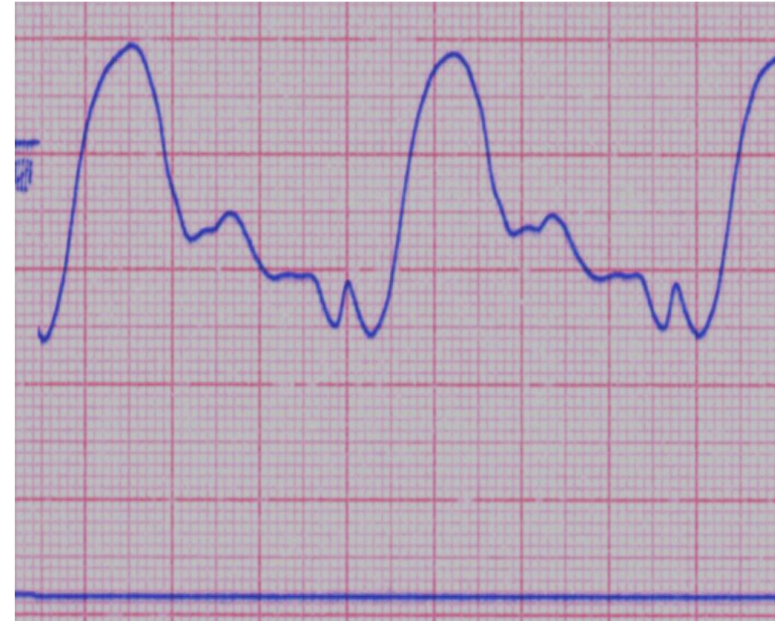
## Doppler echocardiography



## Estimation from TRJV

$$\text{Systolic PAP} = (\text{TRJV max})^2 + \text{RAP(estimated)}$$

## Right Heart Catheterism

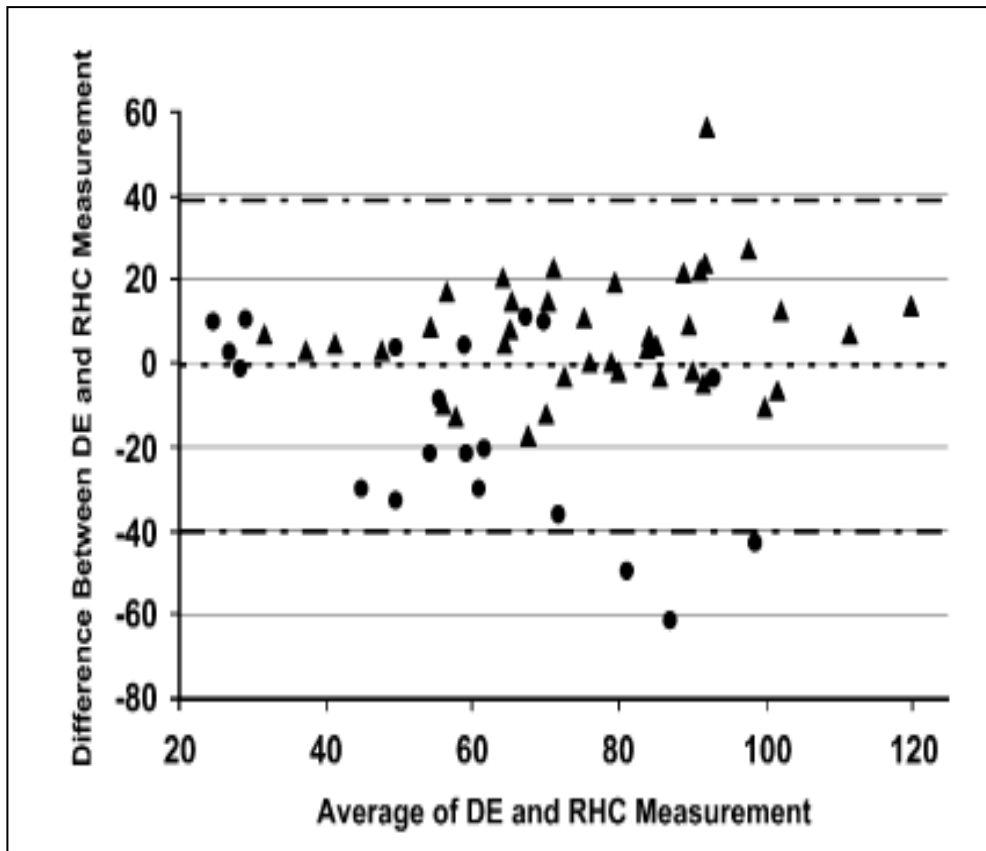


## Gold Standard Tool

## Accuracy of Doppler Echocardiography in the Hemodynamic Assessment of Pulmonary Hypertension

Micah R. Fisher<sup>1\*</sup>, Paul R. Forfia<sup>2†</sup>, Elzbieta Chamera<sup>2</sup>, Traci Houston-Harris<sup>1</sup>, Hunter C. Champion<sup>2</sup>, Reda E. Girgis<sup>1</sup>, Mary C. Corretti<sup>2</sup>, and Paul M. Hassoun<sup>1</sup> **Am J Respir Crit Care Med 2009**

<sup>1</sup>Division of Pulmonary and Critical Care Medicine; <sup>2</sup>Division of Cardiology, Department of Medicine, Johns Hopkins University, Baltimore, Maryland



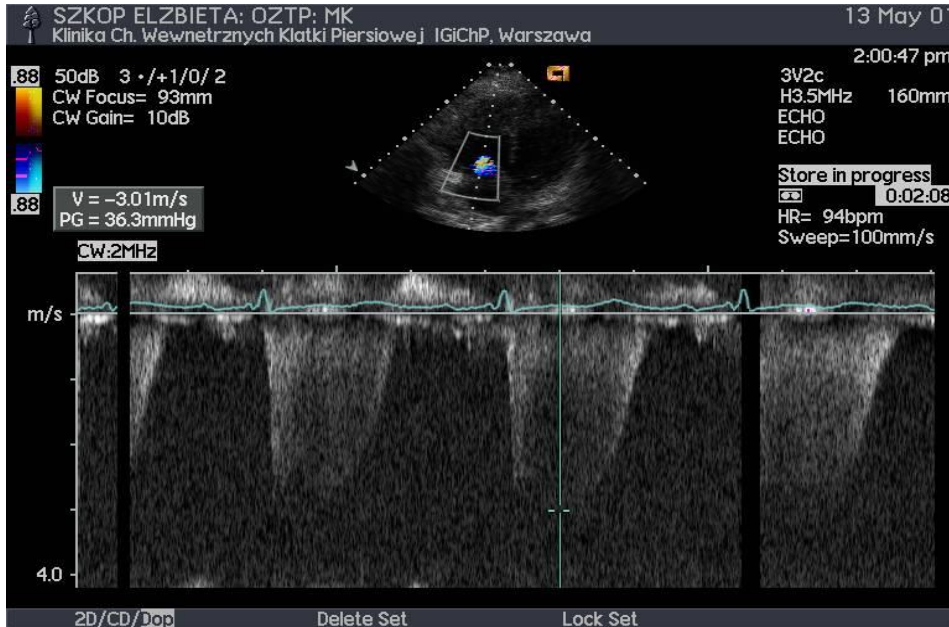
Doppler Echocardiography was inaccurate in 48% of cases ( $\Delta$  greater than  $\pm 10$  mmHg)

Over and under-estimation occurred with a similar frequency (16% vs 15%)



# Measurement of Pulmonary arterial pressure

## Doppler echocardiography

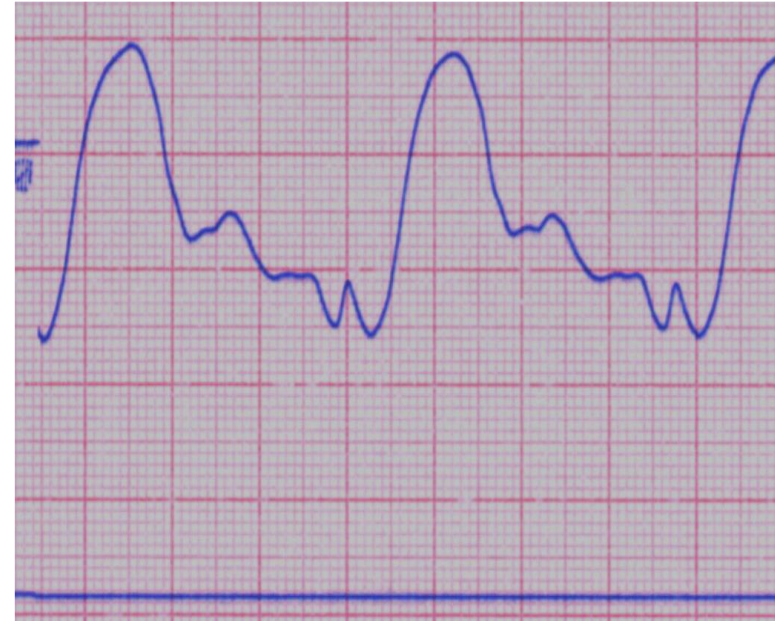


## Estimation from TRJV

Systolic PAP =

$$(\text{TRJV max})^2 + \text{RAP}(\text{estimated})$$

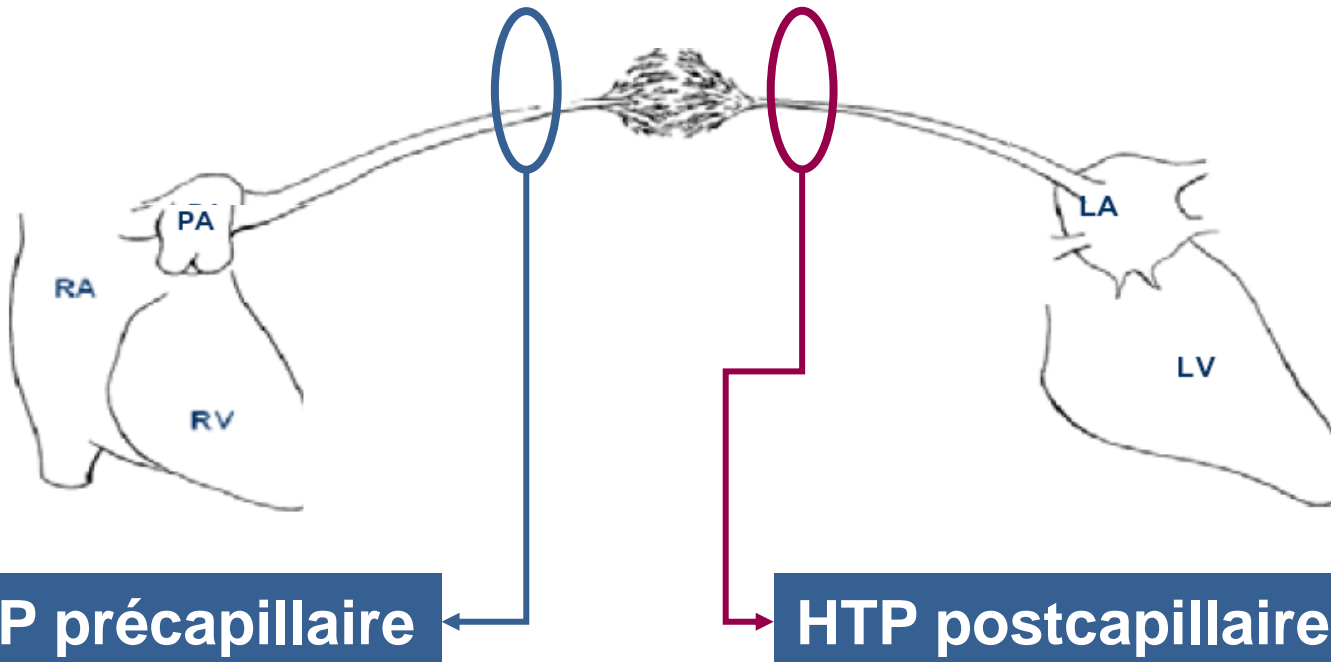
## Right Heart Catheterism



## Gold Standard Tool

# HTP: diagnostic hémodynamique

**PAPm  $\geq 25$  mmHg au repos**



- **PAPm  $\geq 25$  mmHg**
- **PAPO  $\leq 15$  mmHg**
- **DC normal ou abaissé**

- **PAPm  $\geq 25$  mmHg**
- **PAPO  $> 15$  mmHg**
- **DC normal ou abaissé**

# Condensed clinical classification of pulmonary hypertension

1. Pulmonary arterial hypertension (PAH) <b>≈ 15 cas / million*</b>
1.1 Idiopathic <b>≈ 6 cas / million*</b>
1.2 Heritable
1.2.1 BMPR2 mutation
1.2.2 Other mutations
1.3 Drugs and toxins induced
1.4 Associated with:
1.4.1 Connective tissue disease
1.4.2 HIV infection
1.4.3 Portal hypertension
1.4.4 Congenital heart disease (Table 6)
1.4.5 Schistosomiasis
1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis
1". Persistent pulmonary hypertension of the newborn

\* prévalence



European Heart Journal  
doi:10.1093/eurheartj/ehv317

**ESC/ERS GUIDELINES**



# Condensed clinical classification of pulmonary hypertension

<b>1. Pulmonary arterial hypertension (PAH) <math>\approx 15</math> cas / million*</b>
1.1 Idiopathic $\approx 6$ cas / million*
1.2 Heritable <ul style="list-style-type: none"><li>1.2.1 BMPR2 mutation</li><li>1.2.2 Other mutations</li></ul>
1.3 Drugs and toxins induced
1.4 Associated with: <ul style="list-style-type: none"><li>1.4.1 Connective tissue disease</li><li>1.4.2 HIV infection</li><li>1.4.3 Portal hypertension</li><li>1.4.4 Congenital heart disease (Table 6)</li><li>1.4.5 Schistosomiasis</li></ul>
1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis
1". Persistent pulmonary hypertension of the newborn
<b>2. Pulmonary hypertension due to left heart disease</b>
2.1 Left ventricular systolic dysfunction
2.2 Left ventricular diastolic dysfunction
2.3 Valvular disease
2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
2.5 Other

\* prévalence



European Heart Journal  
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**ESC/ERS GUIDELINES**





# Condensed clinical classification of pulmonary hypertension

<b>1. Pulmonary arterial hypertension (PAH) <math>\approx 15</math> cas / million*</b>	<b>3. Pulmonary hypertension due to lung diseases and/or hypoxia</b>
1.1 Idiopathic $\approx 6$ cas / million* 1.2 Heritable 1.2.1 BMPR2 mutation 1.2.2 Other mutations 1.3 Drugs and toxins induced 1.4 Associated with: 1.4.1 Connective tissue disease 1.4.2 HIV infection 1.4.3 Portal hypertension 1.4.4 Congenital heart disease (Table 6) 1.4.5 Schistosomiasis	3.1 Chronic obstructive pulmonary disease 3.2 Interstitial lung disease 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern 3.4 Sleep-disordered breathing 3.5 Alveolar hypoventilation disorders 3.6 Chronic exposure to high altitude 3.7 Developmental lung diseases (Web Table III)
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# Condensed clinical classification of pulmonary hypertension

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<p>1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis</p>	<p><b>4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions</b></p>
<p>1". Persistent pulmonary hypertension of the newborn</p>	<p>4.1 Chronic thromboembolic pulmonary hypertension <math>\approx 3</math> cas / million*</p> <p>4.2 Other pulmonary artery obstructions</p>
<p><b>2. Pulmonary hypertension due to left heart disease</b></p> <p>2.1 Left ventricular systolic dysfunction</p> <p>2.2 Left ventricular diastolic dysfunction</p> <p>2.3 Valvular disease</p> <p>2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies</p> <p>2.5 Other</p>	

\* prévalence



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**ESC/ERS GUIDELINES**



EUROPEAN  
RESPIRATORY  
SOCIETY

# Condensed clinical classification of pulmonary hypertension

<p><b>1. Pulmonary arterial hypertension (PAH) <math>\approx 15</math> cas / million*</b></p> <p>1.1 Idiopathic <math>\approx 6</math> cas / million*</p> <p>1.2 Heritable</p> <p>1.2.1 BMPR2 mutation</p> <p>1.2.2 Other mutations</p> <p>1.3 Drugs and toxins induced</p> <p>1.4 Associated with:</p> <p>1.4.1 Connective tissue disease</p> <p>1.4.2 HIV infection</p> <p>1.4.3 Portal hypertension</p> <p>1.4.4 Congenital heart disease (Table 6)</p> <p>1.4.5 Schistosomiasis</p>	<p><b>3. Pulmonary hypertension due to lung diseases and/or hypoxia</b></p> <p>3.1 Chronic obstructive pulmonary disease</p> <p>3.2 Interstitial lung disease</p> <p>3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern</p> <p>3.4 Sleep-disordered breathing</p> <p>3.5 Alveolar hypoventilation disorders</p> <p>3.6 Chronic exposure to high altitude</p> <p>3.7 Developmental lung diseases (Web Table III)</p>
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<p>1". Persistent pulmonary hypertension of the newborn</p>	<p>4.1 Chronic thromboembolic pulmonary hypertension <math>\approx 3</math> cas / million*</p> <p>4.2 Other pulmonary artery obstructions</p>
<p><b>2. Pulmonary hypertension due to left heart disease</b></p> <p>2.1 Left ventricular systolic dysfunction</p> <p>2.2 Left ventricular diastolic dysfunction</p> <p>2.3 Valvular disease</p> <p>2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies</p> <p>2.5 Other</p>	<p><b>5. Pulmonary hypertension with unclear and/or multifactorial mechanisms</b></p> <p>5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy</p> <p>5.2 Systemic disorders, sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis</p> <p>5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders</p> <p>5.4 Others: pulmonary tumoral thrombotic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension</p>

\* prévalence



**Table 5** Important pathophysiological and clinical definitions

1. Pulmonary hypertension (PH) is a haemodynamic and pathophysiological condition defined as an increase in mean pulmonary arterial pressure  $\geq 25$  mmHg at rest as assessed by right heart catheterization (Table 3). PH can be found in multiple clinical conditions (Table 4).

2. Pulmonary arterial hypertension (PAH, group I) is a clinical condition characterized by the presence of pre-capillary PH (Table 3) and pulmonary vascular resistance  $> 3$  Wood units, in the absence of other causes of pre-capillary PH such as PH due to lung diseases, chronic thromboembolic PH, or other rare diseases (Table 4). PAH includes different forms that share a similar clinical picture and virtually identical pathological changes of the lung microcirculation (Table 4).

3. There is no sufficient data to support the definition of 'PH on exercise'.

Hypertension pulmonaire (HTP):  
PAPm  $\geq 25$  mmHg

Hypertension artérielle pulmonaire (HTAP):

- PAPm  $\geq 25$  mmHg
- PAPO  $\leq 15$  mmHg
- RVP  $> 3$  UW
- Absence d'autres causes d'HTP (PE, maladies pulm)

**Table 8A** Echocardiographic probability of pulmonary hypertension in symptomatic patients with a suspicion of pulmonary hypertension

Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs' <sup>a</sup>	Echocardiographic probability of pulmonary hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

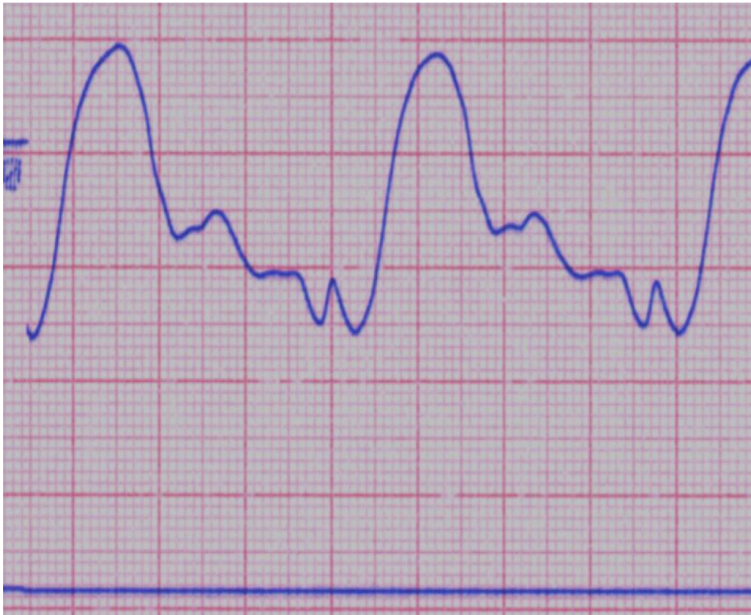
**Table 8B** Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles <sup>a</sup>	B: Pulmonary artery <sup>a</sup>	C: Inferior vena cava and right atrium <sup>a</sup>
Right ventricle/left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior vena cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm <sup>2</sup>
	PA diameter >25 mm.	



# Measurement of Pulmonary arterial pressure

## Right Heart Catheterism



## Gold Standard Tool

- Centres experts
- Morbi-mortalité faible dans les centres experts (1,1% - 0,055%)
- Obligatoire pour confirmer les HTAP et HTP-PE, et pour mettre en place un traitement spécifique de l'HTP.
- $\pm$  Test de vasodilatation (NO inh) pour les HTAPi, HTAPh et toxiques:
  - $\Rightarrow$  Baisse de la PAPm  $\geq 10$  mm Hg
  - $\Rightarrow$  et PAPm  $\leq 40$  mm Hg
  - $\Rightarrow$  et QC augmenté ou inchangé
- $\pm$  Test de remplissage par 500 mL
- Mesures à l'effort moins bien standardisées
- $RVP (UW) = (PAPm - PAPO) / QC$
- Gradient pulmonaire diastolique = PAPd – PAPO
- Gradient transpulmonaire = PAPm – PAPO.

**Table 9** Diagnostic management suggested according to echocardiographic probability of pulmonary hypertension in patients with symptoms compatible with pulmonary hypertension, with or without risk factors for pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension

Echocardiographic probability of PH	Without risk factors or associated condition for PAH or CTEPH <sup>d</sup>	Class <sup>a</sup>	Level <sup>b</sup>	With risk factors or associated conditions for PAH or CTEPH <sup>c</sup>	Class <sup>a</sup>	Level <sup>b</sup>	Ref <sup>c</sup>
Low	Alternative diagnosis should be considered	IIa	C	Echo follow-up should be considered	IIa	C	
Intermediate	Alternative diagnosis, echo follow-up, should be considered	IIa	C	Further assessment of PH including RHC should be considered <sup>e</sup>	IIa	B	45, 46
	Further investigation of PH may be considered <sup>e</sup>	IIb					
High	Further investigation of PH (including RHC <sup>e</sup> ) is recommended	I	C	Further investigation of PH <sup>e</sup> including RHC is recommended	I	C	

CTEPH = chronic thromboembolic pulmonary hypertension; Echo = echocardiographic; PAH = pulmonary arterial hypertension; PH = pulmonary hypertension; RHC = right heart catheterization.

<sup>a</sup>Class of recommendation.

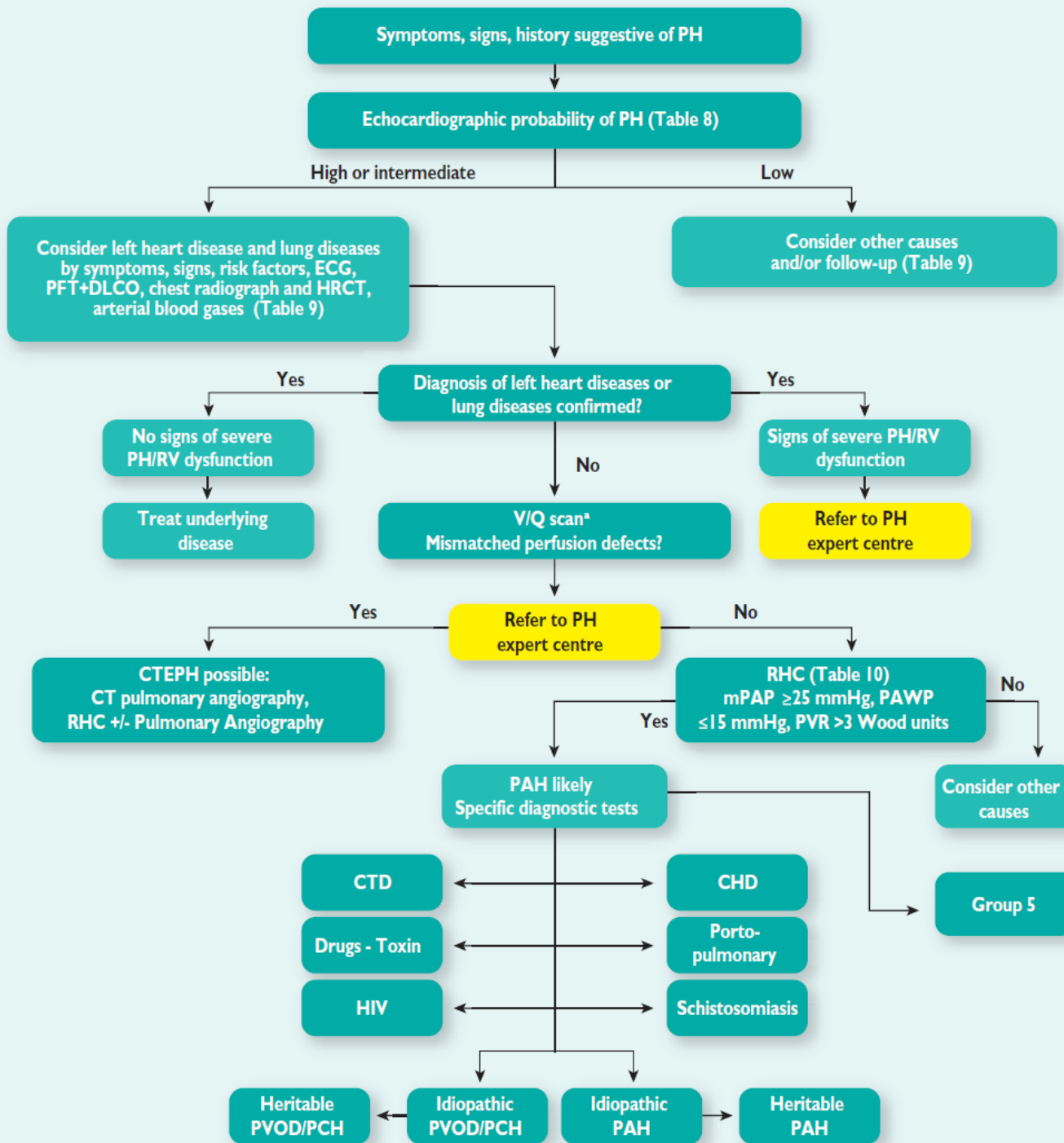
<sup>b</sup>Level of evidence.

<sup>c</sup>Reference(s) supporting recommendations.

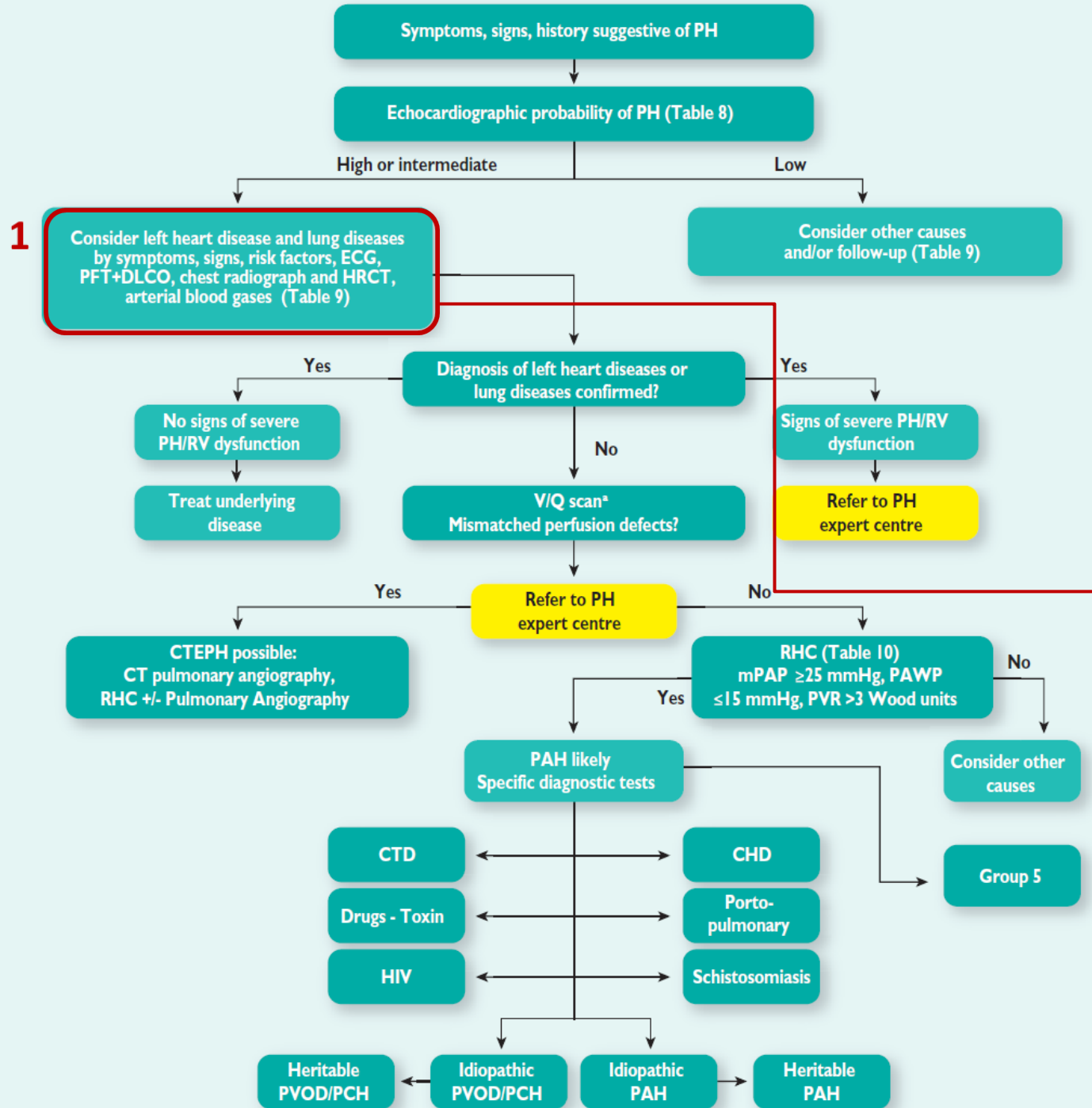
<sup>d</sup>These recommendations do not apply to patients with diffuse parenchymal lung disease or left heart disease.

<sup>e</sup>Depending on the presence of risk factors for PH group 2, 3 or 5.

Further investigation strategy may differ depending on whether risk factors/associated conditions suggest higher probability of PAH or CTEPH – see diagnostic algorithm.

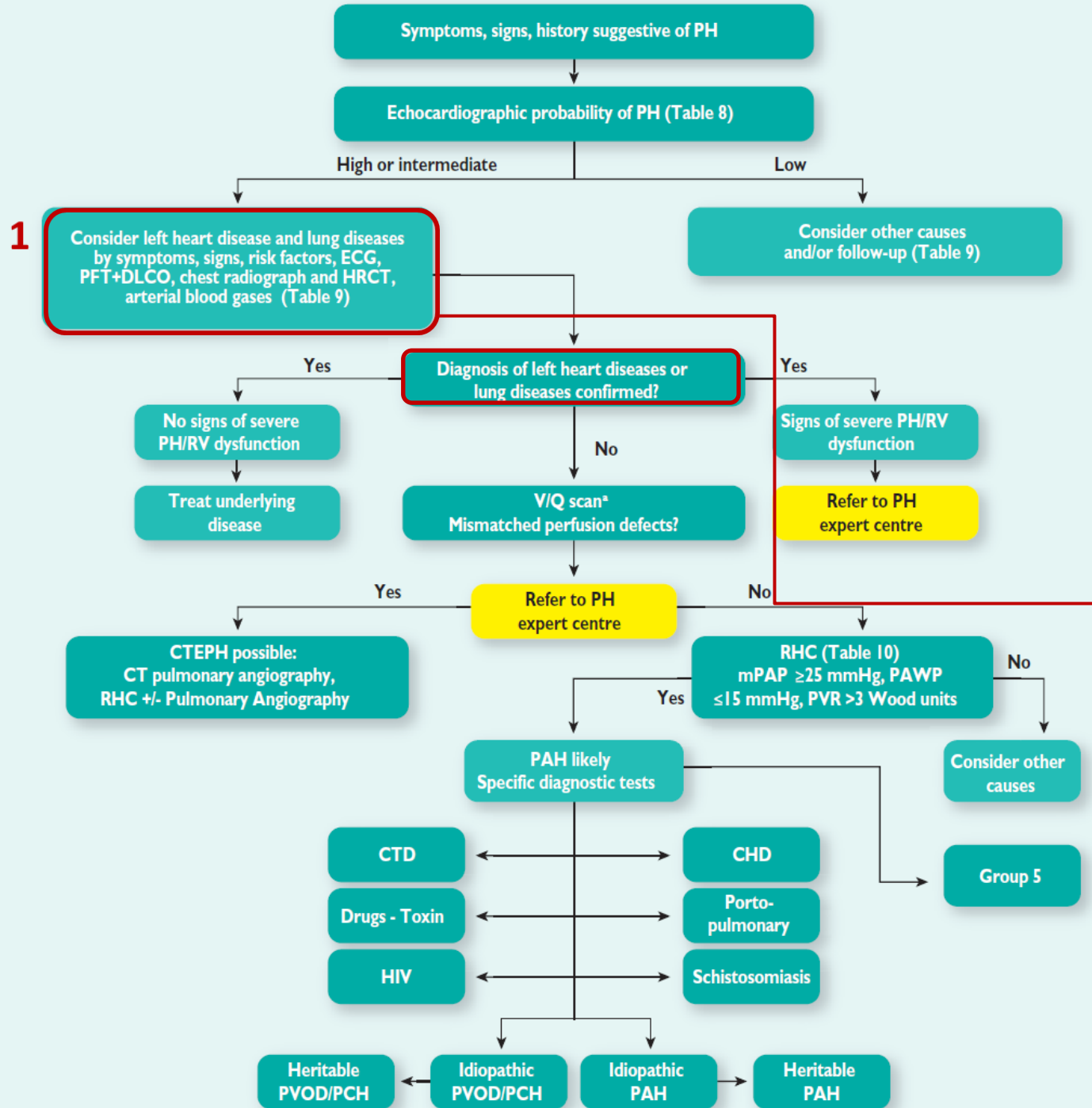


PFT = pulm fonction tests  
 CTD = connective tissue disease  
 CHD = congenital heart diseases  
 PVOD = pulmonary veno-occlusive disease  
 PCH = pulmonary capillary hemangiomathosis



- Clinique
- ECG, Rx thorax
- EFR + DLCO + GDS
- Angioscanner thoracique
- ± Scintigraphie

PFT = pulm fonction tests  
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PFT = pulm fonction tests  
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 PCH = pulmonary capillary hemangiomatosis



# Pulmonary hypertension due to lung disease

## 3. Pulmonary hypertension due to lung diseases and/or hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental lung diseases (Web Table III)

**Table 33** Recommendations for pulmonary hypertension due to lung diseases

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>	Ref. <sup>c</sup>
<u>Echocardiography</u> is recommended for the non-invasive diagnostic assessment of suspected PH in patients with lung disease	I	C	403, 405
<u>Referral to an expert centre</u> is recommended <sup>d</sup> in patients with echocardiographic <u>signs of severe PH</u> and/or severe right ventricular dysfunction	I	C	
<u>The optimal treatment</u> of the underlying lung disease, including long-term O <sub>2</sub> therapy in patients with chronic hypoxaemia, is recommended in patients with PH due to lung diseases	I	C	169
<u>Referral to PH expert center</u> should be considered for patients with signs of severe PH/severe RV failure for individual-based treatment	IIa	C	
RHC is not recommended for suspected PH in patients with lung disease, unless therapeutic consequences are to be expected (e.g. <u>lung transplantation</u> , <u>alternative diagnoses</u> such as PAH or CTEPH, potential enrolment in a clinical trial)	III	C	169
The use of drugs approved for PAH is not recommended in patients with PH due to lung diseases	III	C	411–416

# Pulmonary hypertension due to lung disease

**Table 32** Haemodynamic classification of pulmonary hypertension due to lung disease<sup>9</sup>

Terminology	Haemodynamics (right heart catheterization)
COPD/IPF/CPFE without PH	PAPm <25 mmHg
COPD/IPF/CPFE with PH	PAPm ≥25 mmHg
COPD/IPF/CPFE with severe PH	PAPm >35 mmHg, or PAPm ≥25 mmHg in the presence of a low cardiac output (CI <2.5 L/min, not explained by other causes)

CI = cardiac index; COPD = chronic obstructive pulmonary disease; CPFE = combined pulmonary fibrosis and emphysema; IPF = idiopathic pulmonary fibrosis; PAP = pulmonary artery pressure; PAPm = mean pulmonary arterial pressure; PH = pulmonary hypertension.

Penser à l'association avec

- Cardiopathie gauche
- HTP PE et MTEV
- ± association HTAP



## Indications du cathétérisme cardiaque droit:

- Doit être réalisé dans des centres experts
- Confirmation du diagnostic de l'HTAP, décision thérapeutique et évaluation de l'efficacité
- Cardiopathie congénitales
- En cas de cardiopathie gauche suspectée ou associée pour le diagnostic différentiel avec cardiopathie gauche et décision thérapeutique
- En cas de maladie pulmonaire chronique:
  - ⇒ si une transplantation est discutée
  - ⇒ Ou pour le diagnostic différentiel avec cardiopathie gauche et décision thérapeutique (HTP sévères)
- En cas d'hypertension pulmonaire post-embolique

# Pulmonary hypertension due to lung disease

**Table 2** Management of PH in the Setting of Chronic Lung Disease

Underlying Lung Disease	mPAP <25 mm Hg at Rest	mPAP ≥25 and <35 mm Hg at Rest	mPAP ≥35 mm Hg at Rest*
COPD with FEV1 ≥60% of predicted IPF with FVC ≥70% of predicted CT: absence of or only very modest airway or parenchymal abnormalities	No PH No PAH treatment recommended	PH classification uncertain No data currently support treatment with PAH-approved drugs	PH classification uncertain: discrimination between PAH (group 1) with concomitant lung disease or PH caused by lung disease (group 3) Refer to a center with expertise in both PH and chronic lung disease
COPD with FEV1 <60% of predicted IPF with FVC <70% of predicted Combined pulmonary fibrosis and emphysema on CT	No PH No PAH treatment recommended	PH-COPD, PH-IPF, PH-CPFE No data currently support treatment with PAH-approved drugs	Severe PH-COPD, severe PH-IPF, severe PH-CPFE Refer to a center with expertise in both PH and chronic lung disease for individualized patient care because of poor prognosis; randomized controlled trials required

\*Lower PA pressures may be clinically significant in COPD/DPLD patients with depressed cardiac index or right ventricular dysfunction.  
CPFE = combined pulmonary fibrosis and emphysema; mPAP = mean pulmonary artery pressure; other abbreviations as in Table 1.

## Traitement :

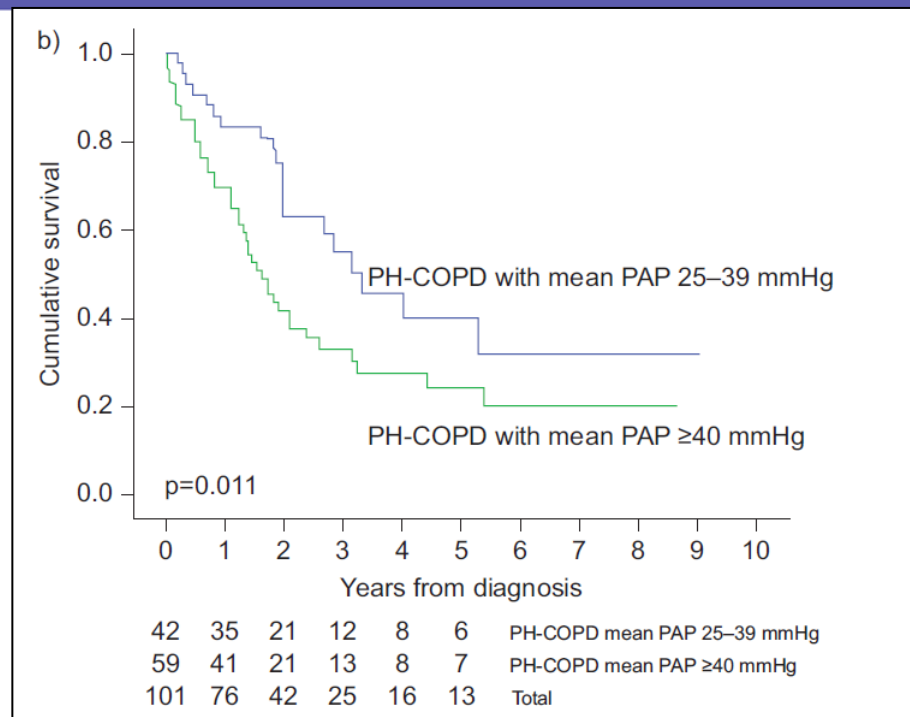
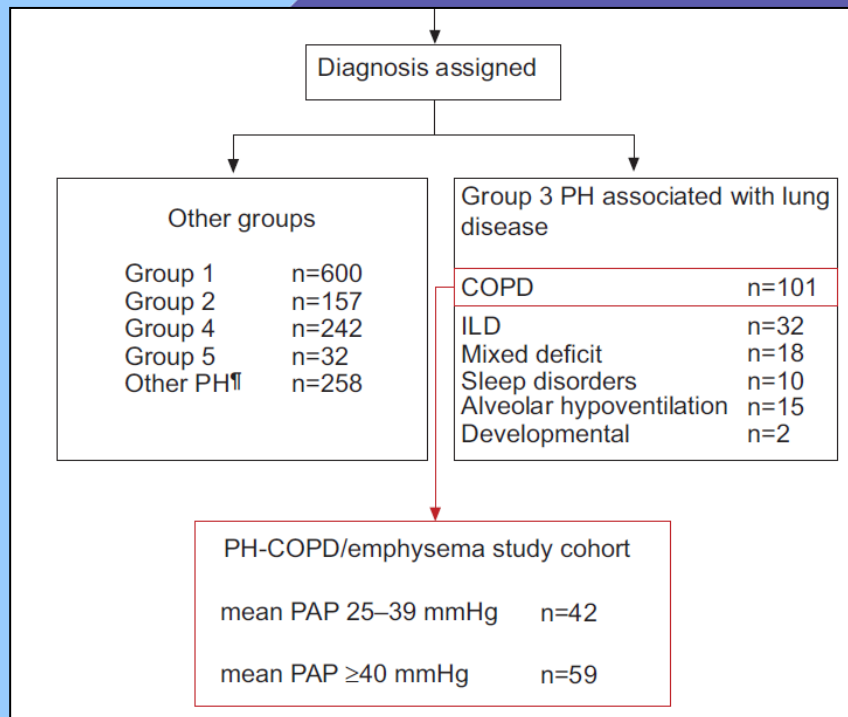
- De la pathologie
- OLD (BPCO)
- traitement vaso-actif: non recommandé (absence d'efficacité; aggravation potentielle des échanges gazeux)
- Si HTP sévère: à discuter dans centre de référence

*Seeger et al. JACC Vol. 62, No. 25, Suppl D, 2013*

## Intérêt d'un registre des patients BPCO + HTP « sévère »

- Registre PHRC Hervé Mal
- Détecter les pts BPCO avec maladie vasculaire prédominante
- Association fortuite BPCO - HTAP ou vraie HTP liée à la BPCO?
- Décrire profil hémodynamique, fonctionnel et pronostic
- Facteurs prédisposants
- Evaluer les possibilités thérapeutiques

# PH in COPD: results from the ASPIRE registry



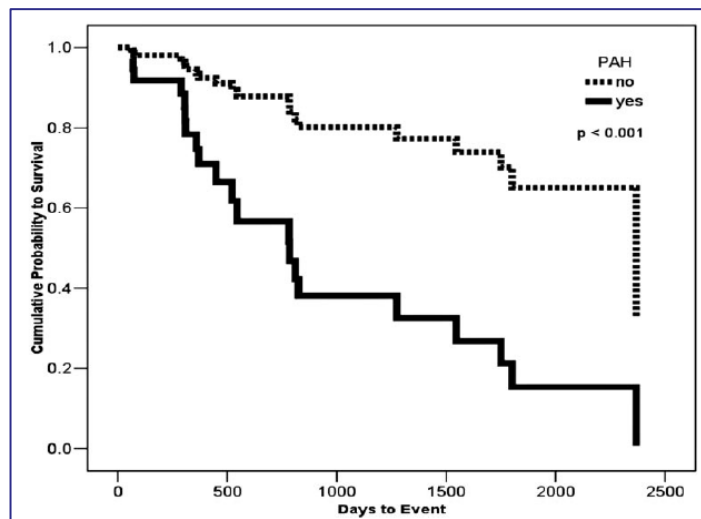
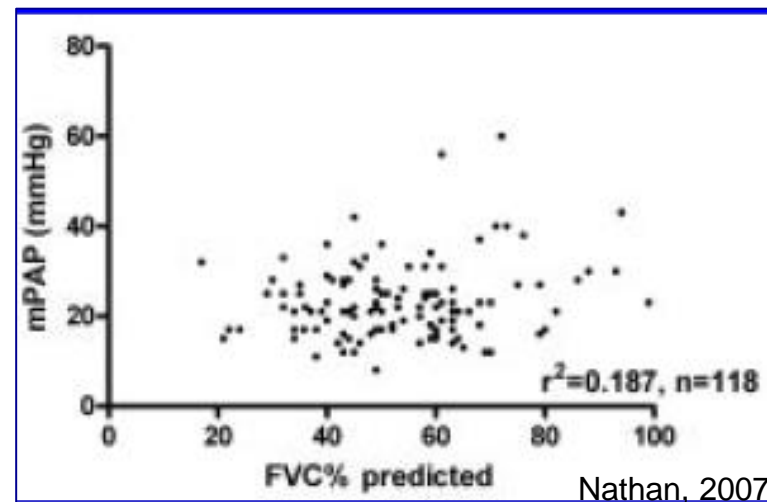
**TABLE 1** Baseline characteristics

	COPD mean PAP 25–39 mmHg	COPD mean PAP ≥40 mmHg	p-value
Subjects n	42	59	
Age years	67 ± 11	70 ± 9	0.092
<b>Pulmonary function tests</b>			
FEV <sub>1</sub> % pred	51 ± 28	65 ± 23	0.006
FVC % pred	78 ± 25	90 ± 24	0.022
FEV <sub>1</sub> /FVC	0.51 ± 0.18	0.59 ± 0.18	0.041
DLCO % pred	40 ± 20	27 ± 13	0.001
Mean RAP mmHg	8 ± 4	12 ± 5	0.001
Mean PAP mmHg	32 ± 5	49 ± 8	<0.001
CI L · min <sup>-2</sup>	3.2 ± 0.8	2.5 ± 0.7	<0.001
PCWP mmHg	13 ± 5	12 ± 5	0.156
PVR dyn · s · cm <sup>-5</sup>	303 ± 168	755 ± 377	<0.001
SvO <sub>2</sub> %	67 ± 8	63 ± 8	0.051



# HTP et Pneumopathies interstitielles

- Absence de corrélation de l'HTP avec les paramètres fonctionnels respiratoires.



## 79 FPI, bilan pré TP

- CV  $\approx$  50% th
- PAPm =  $29.5 \pm 3.3$  mmHg
- HTP chez 32% des patients
- Mortalité à 1 an: 28% si HTP vs 5.5% sans HTP ( $p=0.002$ )

*Simonneau G et al, J Heart Lung Transplant 1997;16:460*

*Lettieri CJ, Chest 2006;129:746 ;*

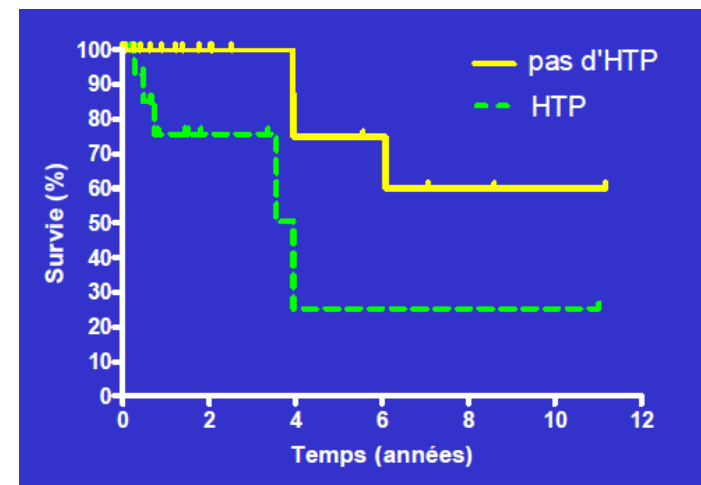
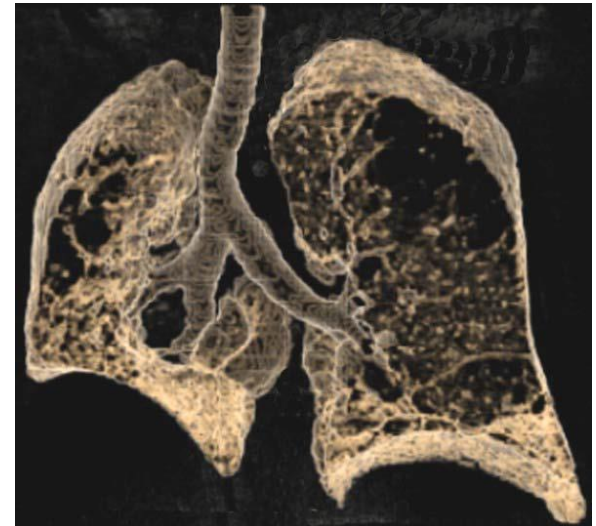
*Nathan SD Chest 2007;131:657 ;*

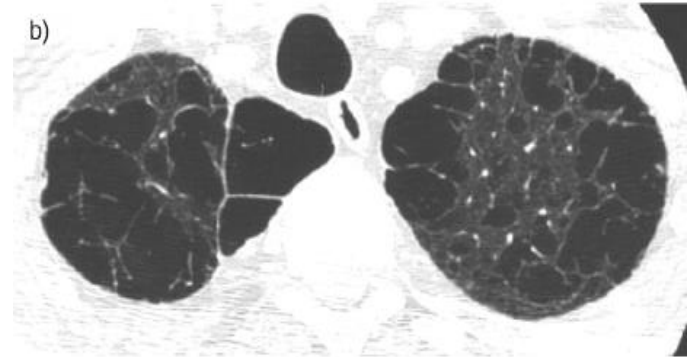
*Harari S, Shorr AF Eur Respir J 2007;30:715)*

# Syndrome d'emphysème et fibrose pulmonaire combinés

*Cottin V et al, et le GERMOP, Eur Respir J 2005;26:586*

- Tabagisme constant
- Dyspnée d'effort importante mais spirométrie subnormale
- Altération majeure des échanges gazeux (DLco, désaturation d'exercice)
- Diagnostic par TDM (emphysème paraseptal)
- Hypertension pulmonaire fréquente (~50%) et de mauvais pronostic





*V Cottin et al, Eur Resp J 2005*

# Sarcoïdose et HTP

- Prévalence de l'HTP associée aux sarcoïdoses estimée à 5,4% <sup>1</sup>
  - Plus fréquente dans les formes fibrosantes <sup>2,3</sup>
- Mécanismes multifactoriels
  - Groupe V des HTP <sup>4</sup>
- Impact sur le pronostic
  - Mortalité multipliée par 8 <sup>5</sup>

<sup>1</sup> Handa, Chest 2006; 129:1246–1252

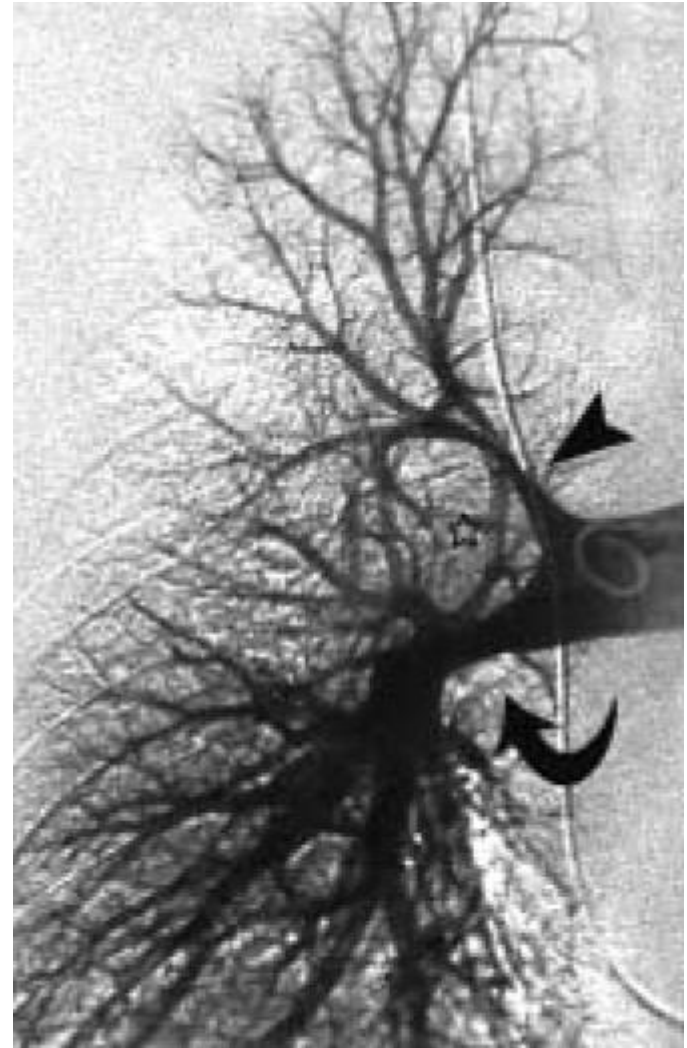
<sup>2</sup> Baughman, Chest 2010; 138:1078–1085

<sup>3</sup> Sulica, Chest 2005;128:1483-1489

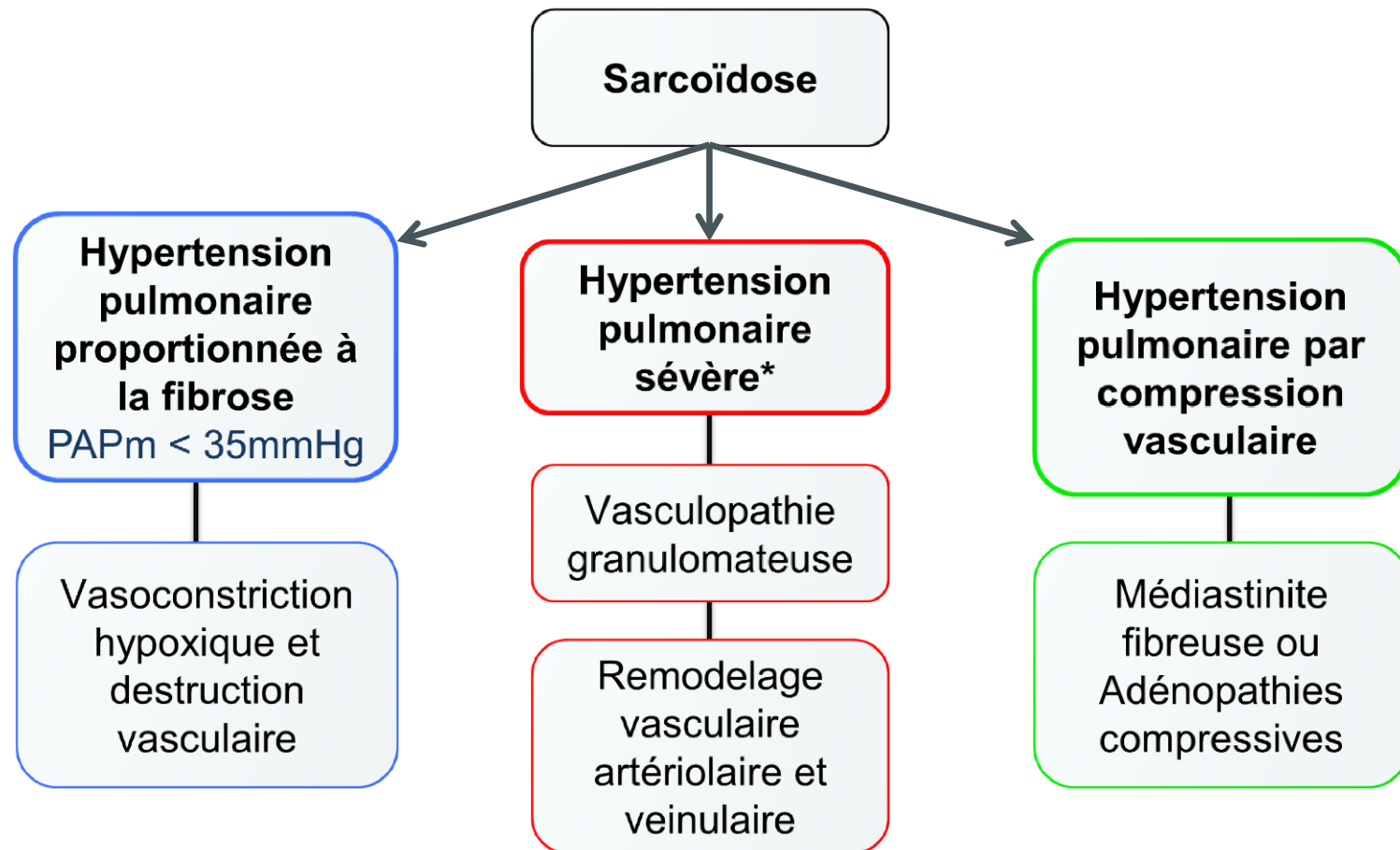
<sup>4</sup> ESC/ERS Guidelines, Eur Respir J 2015

<sup>5</sup> Nardi, Eur Respir J 2011; 38: 1368–1373

# HTP et sarcoïdose



# Mécanismes des HTP précapillaires



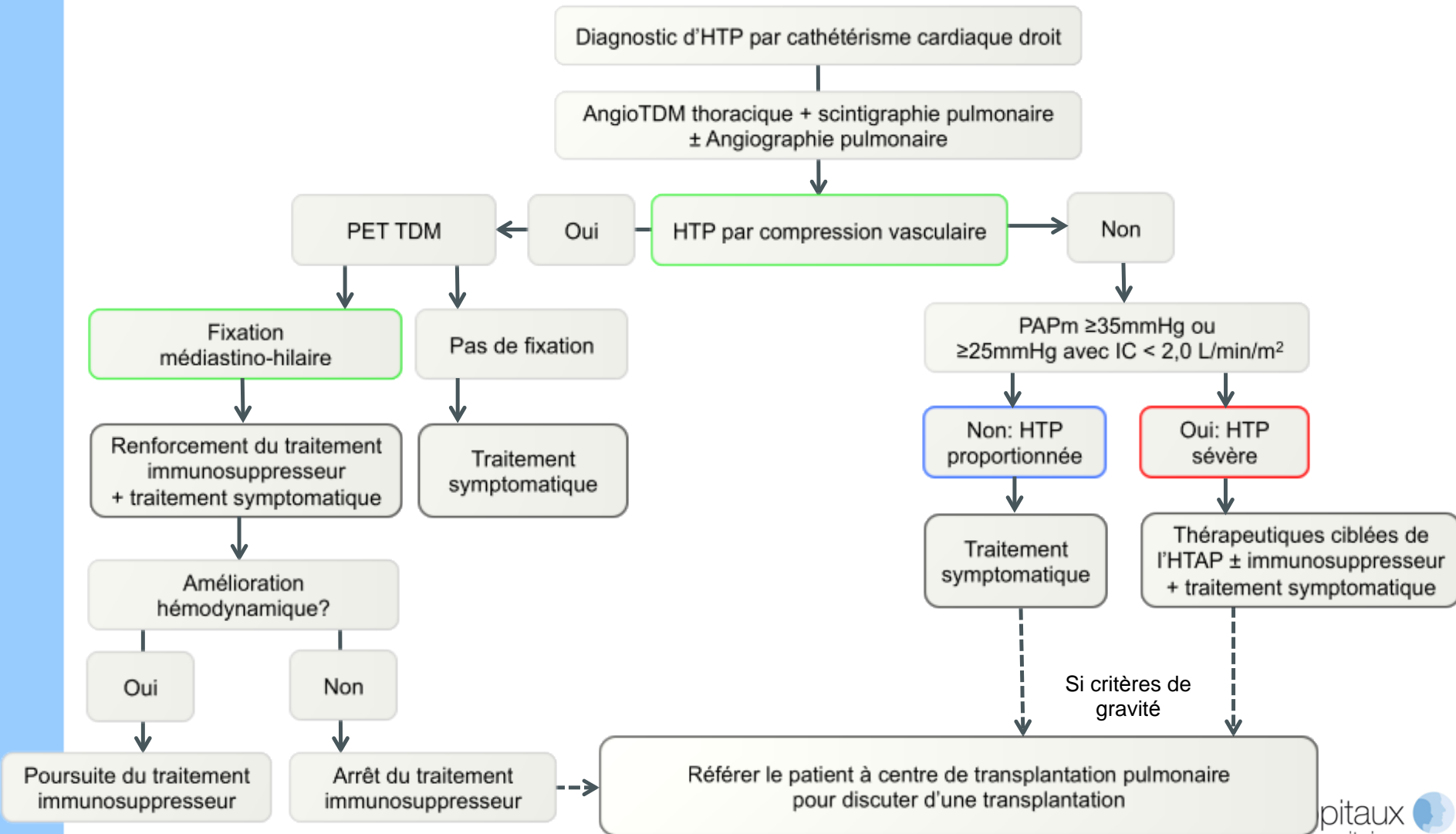
\*PAPm  $\geq$  35mmHg ou PAPm  $\geq$  25mmHg et un IC  $<$  2,0 L/min/m<sup>2</sup>



# Traitement: HTP et sarcoïdose

- Les thérapeutiques ciblées de l'HTAP
  - Améliorent l'hémodynamique des HTP sévères
  - Sans amélioration du TM6
  - Pas d'efficacité dans les autres groupes
- Les corticoïdes/immunosuppresseurs:
  - Semblent efficaces dans les formes « actives » d'HTP par compression vasculaire
  - Efficaces dans certaines formes d'HTP sévère?
- Le pronostic à long terme reste sombre
  - Mortalité à 5 ans proche de 50%
- Prise en charge dans centres experts:
  - Evaluation hémodynamique
  - Bilan complet: mécanisme

# Algorithme de prise en charge



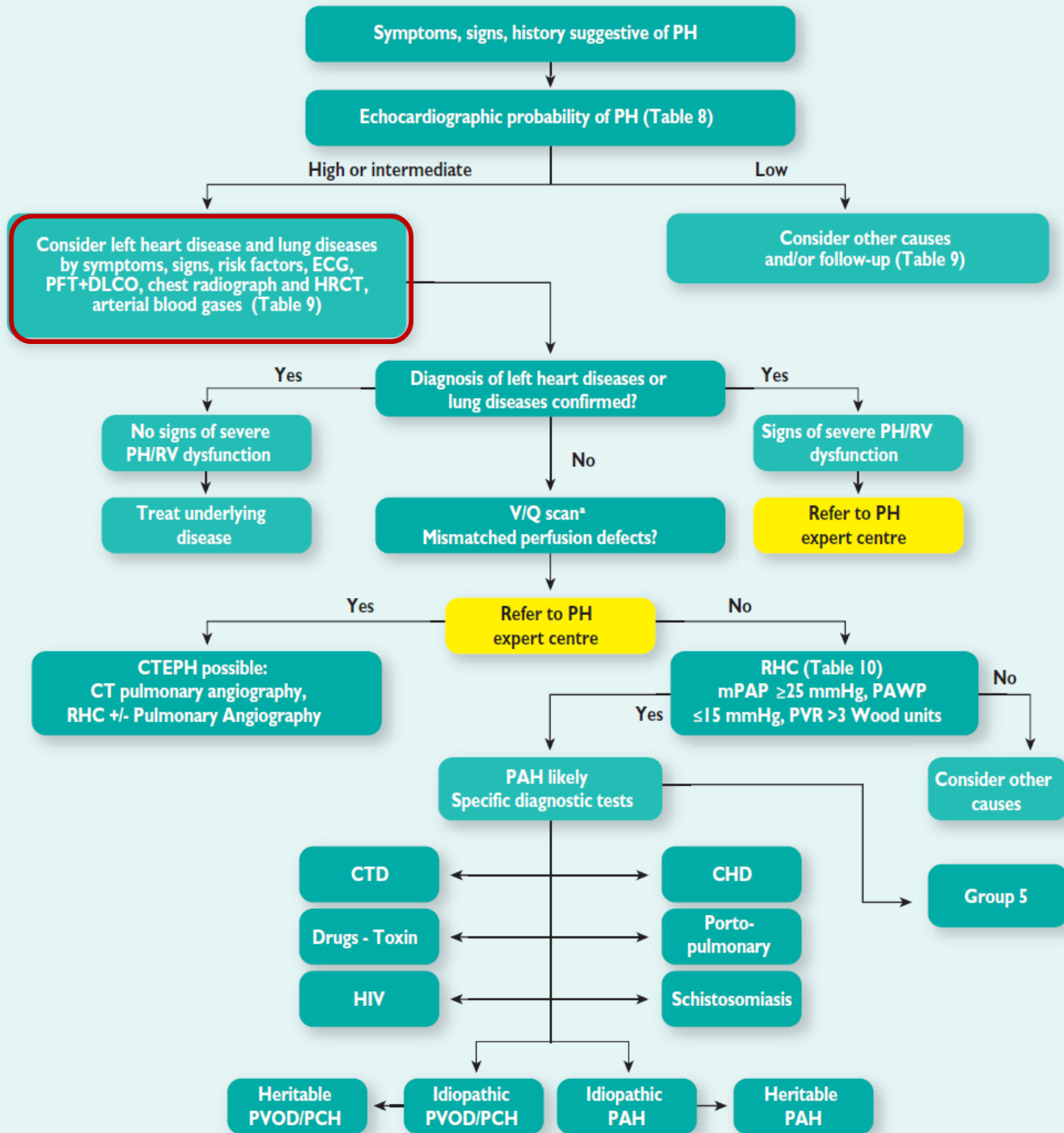
# Résultats: Traitements reçus

- **Thérapeutique ciblée de l'HTAP, n=107**
  - Monothérapies orales (68 ARE, 21 IPDE5), n=89
  - Bithérapies orales (ARE + IPDE5), n=12
  - Bithérapies (ARE ou IDE5 + prostacycline IV SC ou inhalée), n=4
  - Prostacycline IV, n=1
  - Prostacycline inhalée, n=1
- **Corticothérapie ou immunosuppresseurs, n=34**
  - Dont 16 ayant une thérapeutique ciblée HTAP associée
  - Corticothérapie ou immunosuppresseurs seuls, n=18
- **Aucun traitement, n=31**

# Effet à court terme des thérapeutiques ciblées de l'HTP

HTP SEVERE				
n= 69	Bilan initial	4 – 6 mois	Différence	p
<b>NYHA I-II/III/IV</b>	8 / 45 / 16	24 / 36 / 9		0,004
<b>TM6 (mètres)</b>	323 (±116)	331 (±132)	+ 8m	0,58
<b>POD (mmHg)</b>	8 (±4)	6 (±5)	- 18%	0,014
<b>PAPm (mmHg)</b>	49 (±9)	43 (±11)	- 11%	<0,00008
<b>IC (L/min/m<sup>2</sup>)</b>	2,6 (±0,8)	2,9 (±0,8)	+12%	<0,00001
<b>RVP (UW)</b>	9,7 (±4,3)	6,9 (±2,9)	- 29%	<0,00001
HTP PROPORTIONNEE				
n=11	Bilan initial	4 – 6 mois	Différence	p
<b>NYHA I-II/III/IV</b>	3 / 5 / 3	5 / 5 / 1		0,02
<b>TM6 (mètres)</b>	382 (±154)	369 (±192)		0,58
<b>POD (mmHg)</b>	4 (±3)	3 (±3)		0,36
<b>PAPm (mmHg)</b>	32 (±3)	30 (±6)		0,21
<b>IC (L/min/m<sup>2</sup>)</b>	3,0 (±0,7)	3,0 (±1,0)		0,78
<b>RVP (UW)</b>	5,0 (±0,6)	4,7 (±1,5)		0,35

1



PFT = pulm fonction tests  
 CTD = connective tissue disease  
 CHD = congenital heart diseases  
 PVOD = pulmonary veno-occlusive disease  
 PCH = pulmonary capillary hemangiomathosis

**Table 30** Examples of key factors suggestive of group 2 pulmonary hypertension

Clinical presentation	Echocardiography	Other features
Age >65 years	Structural left heart abnormality <ul style="list-style-type: none"> <li>• Disease of left heart valves</li> <li>• LA enlargement (&gt;4.2 cm)</li> <li>• Bowing of the IAS to the right</li> <li>• LV dysfunction</li> <li>• Concentric LV hypertrophy and/or increased LV mass</li> </ul>	ECG <ul style="list-style-type: none"> <li>• LVH and/or LAH</li> <li>• AF/Afib</li> <li>• LBBB</li> <li>• Presence of Q waves</li> </ul>
Symptoms of left heart failure	Doppler indices of increased filling pressures <ul style="list-style-type: none"> <li>• Increased E/e'</li> <li>• &gt;Type 2–3 mitral flow abnormality</li> </ul>	Other imaging <ul style="list-style-type: none"> <li>• Kerley B lines</li> <li>• Pleural effusion</li> <li>• Pulmonary oedema</li> <li>• LA enlargement</li> </ul>
Features of metabolic syndrome	Absence of <ul style="list-style-type: none"> <li>• RV dysfunction</li> <li>• Mid systolic notching of the PA flow</li> <li>• Pericardial effusion</li> </ul>	
History of heart disease (past or current)		
Persistent atrial fibrillation		

AF = atrial flutter; Afib = atrial fibrillation; ECG = electrocardiogram; IAS = inter-atrial septum; LA = left atrium; LAH = left atrial hypertrophy/dilatation; LBBB = left bundle branch block; LV = left ventricle; LVH = left ventricular hypertrophy; PA = pulmonary artery; RV = right ventricle.

## Indications du cathétérisme cardiaque droit:

- Doit être réalisé dans des centres experts
- Confirmation du diagnostic de l'HTAP, décision thérapeutique et évaluation de l'efficacité
- Cardiopathie congénitales
- En cas de cardiopathie gauche suspectée ou associée : diagnostic différentiel, HTP mixte, décision thérapeutique
- En cas de maladie pulmonaire chronique:
  - ⇒ si une transplantation est discutée
  - ⇒ Ou pour le diagnostic différentiel avec cardiopathie gauche, HTP mixte et décision thérapeutique
- En cas d'hypertension pulmonaire post-embolique



**Table 3 Haemodynamic definitions of pulmonary hypertension<sup>a</sup>**

Definition	Characteristics <sup>a</sup>	Clinical group(s) <sup>b</sup>
PH	PAPm $\geq$ 25 mmHg	All
Pre-capillary PH	PAPm $\geq$ 25 mmHg PAWP $\leq$ 15 mmHg	1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	PAPm $\geq$ 25 mmHg PAWP $>$ 15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Isolated post-capillary PH (Ipc-PH)	DPG $<$ 7 mmHg and/or PVR $\leq$ 3 WU <sup>c</sup>	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG $\geq$ 7 mmHg and/or PVR $>$ 3 WU <sup>c</sup>	

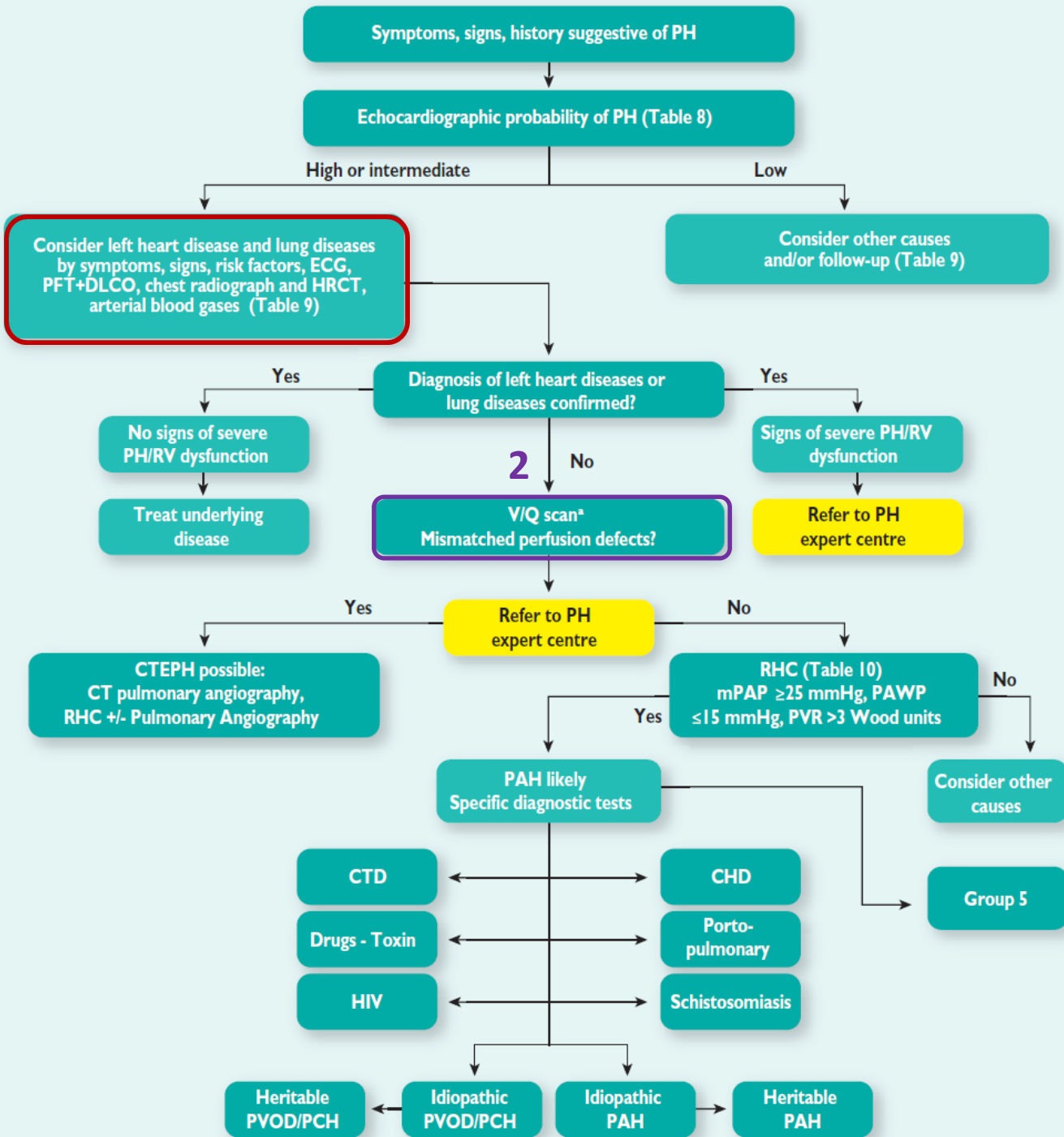
CO = cardiac output; DPG = diastolic pressure gradient (diastolic PAP – mean PAWP); mPAP = mean pulmonary arterial pressure; PAWP = pulmonary arterial wedge pressure; PH = pulmonary hypertension; PVR = pulmonary vascular resistance; WU = Wood units.

<sup>a</sup>All values measured at rest; see also section 8.0.

<sup>b</sup>According to Table 4.

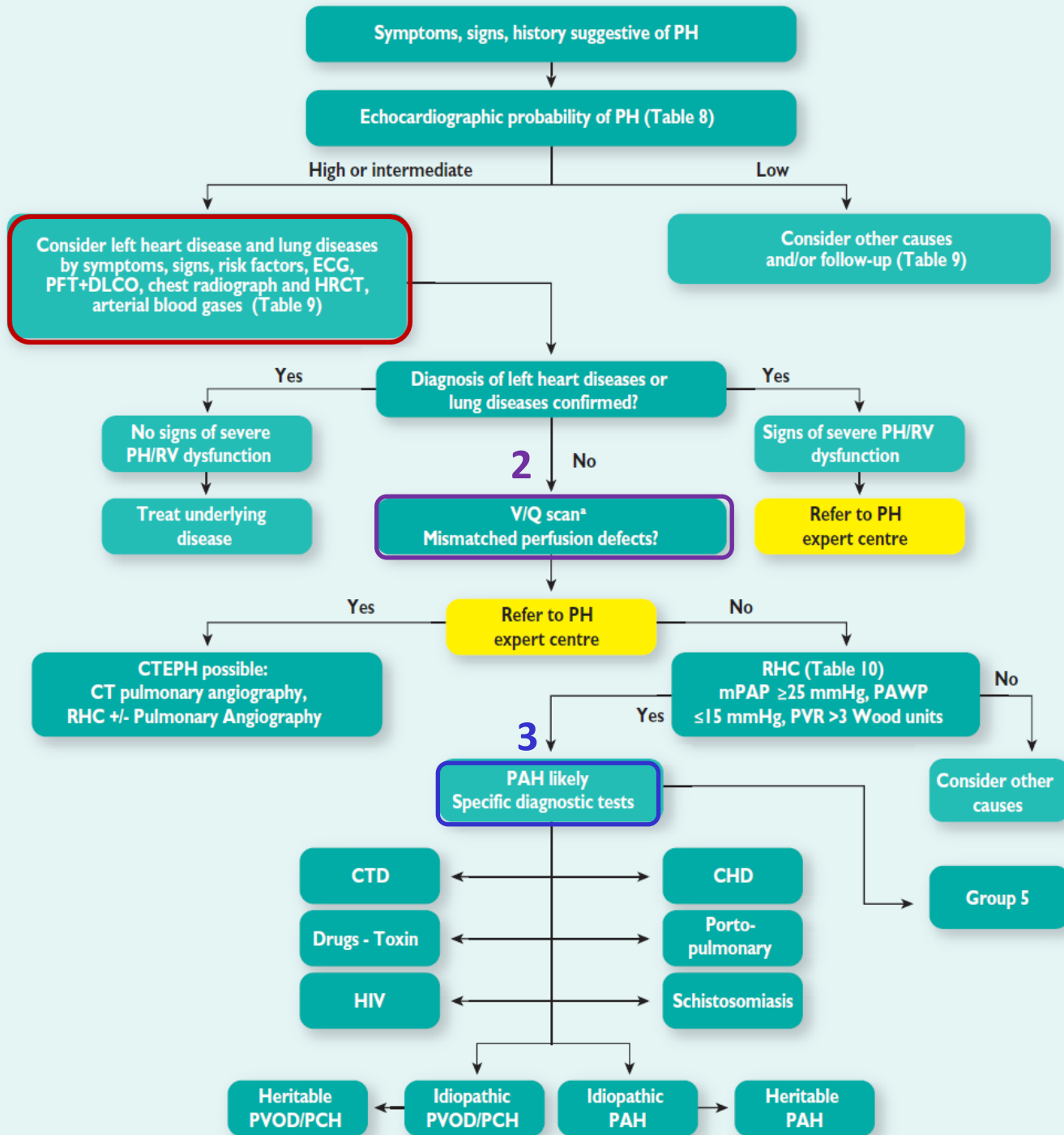
<sup>c</sup>Wood Units are preferred to dynes.s.cm<sup>-5</sup>.

1



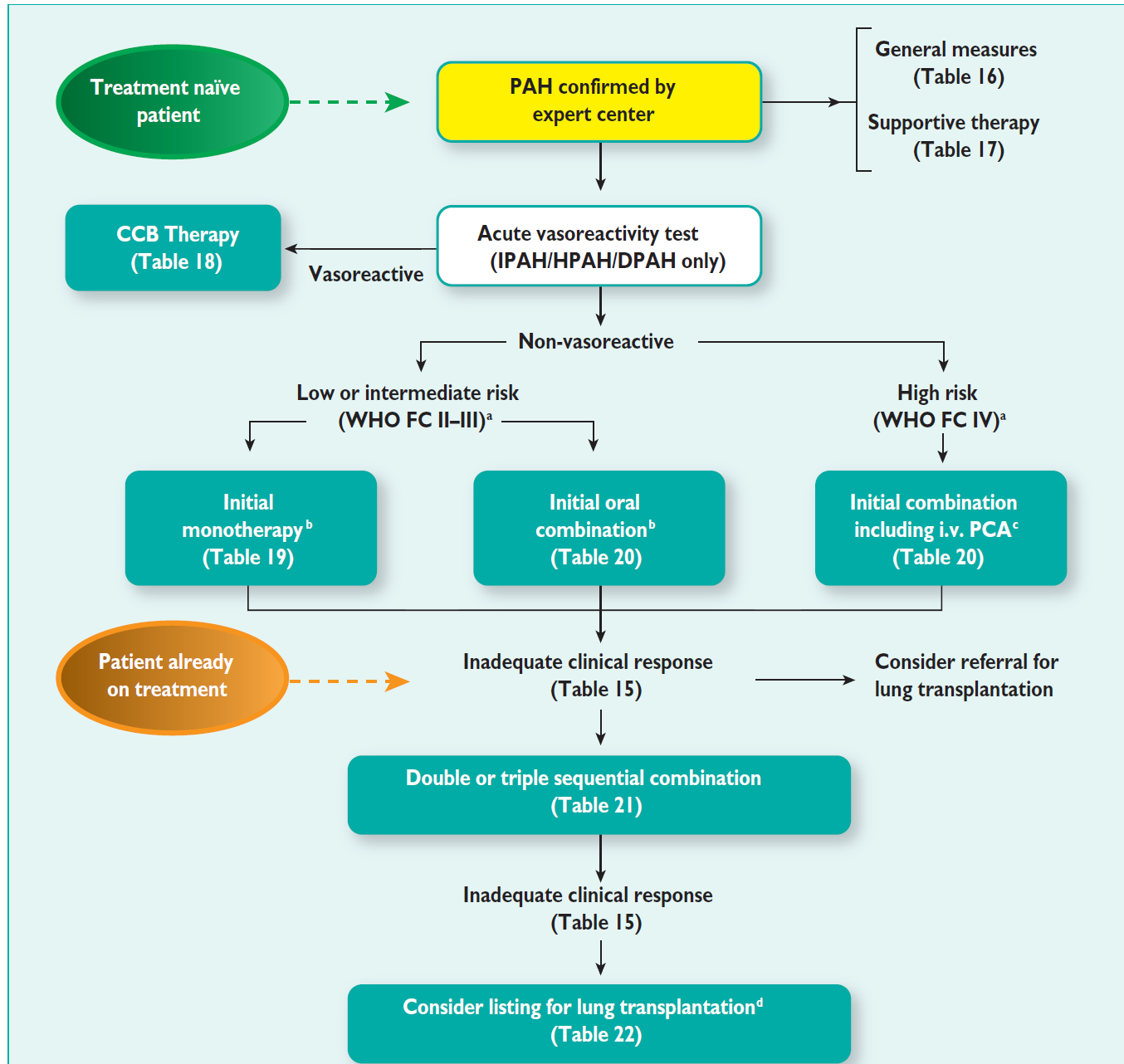
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# Treatment algorithm for pulmonary arterial hypertension patients (group 1)



# Conclusion

- Confirmation par cathétérisme de toute suspicion d'HTTP, mais...
- Rechercher étiologies les plus fréquentes
- Si HTP non sévère des groupes 2 et 3, le cathétérisme n'est pas systématique
- Penser à la MTEV et à l'HTP PE.  
Association
- HTP sévère ou des contextes plus particuliers: centre expert